Experiences of Siblings of Patients With Fanconi Anemia

Sadie P. Hutson, PhD, RN, WHNP[1,2]* and Blanche P. Alter, MD, MPH[2]

Background. Clinical management of families with autosomal recessive genetic disorders focuses almost exclusively on the affected family members. However, clinically unaffected members of such families may also be severely troubled by the serious illness in a family member. The purpose of this study was to explore the experiences of healthy siblings of patients with a chronic genetic disease, Fanconi Anemia (FA). ***Procedure.*** We used a qualitative, descriptive design, which consisted of in-depth, semi-structured interviews. A convenience sample of nine siblings of patients with FA was recruited from a National Cancer Institute clinical research protocol, which targets families with inherited bone marrow failure syndromes. NVivo 2.0 software facilitated qualitative content analysis of the data. ***Results.*** Siblings' rich descriptions provided novel insights into the intricate hardships of living within a family in which a rare, life-threatening, chronic genetic illness in one member is the focus of daily life. Four major themes of the sibling experience emerged from the interview data: (1) containment, (2) invisibility, (3) worry, and (4) despair. ***Conclusions.*** Our data suggest that unrecognized psychosocial issues exist for the apparently healthy siblings of patients with FA. This study explores the psychosocial consequences of living in a family with FA and one of only a few studies to explore the sibling experience of chronic illness using a contemporaneous approach. These findings support the need for an increased awareness among health care providers; future hypothesis driven investigation, and improved assessment of problems with potential psychological morbidity. Pediatr Blood Cancer
Published 2006 Wiley-Liss, Inc.†

Key words: cancer; chronic disease; fanconi anemia; genetics; psychosocial aspects; siblings

INTRODUCTION

Clinical management of high-risk families with autosomal recessive genetic disorders focuses almost exclusively on the affected family members. However, clinically unaffected members of such families may also be severely troubled by the serious illness in a family member. To begin to understand the extent of this problem, we studied the experiences of healthy siblings living with children with Fanconi Anemia (FA), a complex, rare autosomal recessive genetic disease with extremely high rates of aplastic anemia, acute myelogenous leukemia, and solid tumors [1]. The syndrome includes a broad array of congenital and developmental abnormalities, with the potential for affecting nearly every organ system in the body. Consequently, the diagnosis of FA presents an enormous challenge to patients, otherwise healthy siblings, and parents.

During normal growth and development, interactions with parents and siblings in the context of family life influence and shape a child's personality and intellect. Although the developmental importance of the sibling relationship has been widely recognized [2–5], most current research on sibling relationships focuses on adult life, rather than the experiences in childhood upon which the later relationship is based.

The diagnosis of serious childhood illness places extreme demands on all members of the family. Major themes in the literature center on the negative experiences of siblings such as loss of attention, loss of the family's routines, loss of security, loss of companionship of the ill child, loneliness, confusion, guilt, fear, jealousy, increased responsibility, sadness, anxiety, and embarrassment [6–19]. Positive aspects have also been reported, such as gains in maturity, understanding, compassion, improved self-confidence, increased empathy for parents, and closer family bonds [12,14–16,20–23].

While the benefits of psychosocial support for patients and families with a genetic predisposition to cancer and chronic illness have been widely assumed, there is little published information regarding the identification and range of psychosocial issues or possible interventions for meeting these psychosocial needs. In particular, the psychosocial consequences for children living in a family with FA have not been described.

Much of the research on siblings of chronically ill children has targeted childhood cancer [6,9–12,14–17,19,21,22,24–33]. Most of these data were collected by self-report questionnaires and focused on coping strategies, stressors, and family adaptation. A missing link in prior research is the perspective and voice of the healthy sibling, including contemporaneous rather than retrospective reports of their experiences.

[1]Department of Family/Community Nursing, College of Nursing, East Tennessee State University, Tennessee; [2]Clinical Genetics Branch, Division of Cancer Epidemiology and Genetics, National Cancer Institute, National Institutes of Health, Dept. of Health and Human Services, Rockville, Maryland

Grant sponsor: National Cancer Institute Cancer Research Training Award (to S.P.H.); Grant sponsor: National Institute of Nursing Research T32 Pre-doctoral Training Award in Psychosocial Oncology (University Of Pennsylvania School Of Nursing) (to S.P.H.); Grant sponsor: American Cancer Society Doctoral Degree Scholarship in Cancer Nursing (to S.P.H.); Grant sponsor: Virginia Henderson Clinical Research Award, Sigma Theta Tau International (to S.P.H.).

*Correspondence to: Sadie P. Hutson, East Tennessee State University, College of Nursing, Room 254, PO Box 70676, Johnson City, TN 37614. E-mail: hutsons@mail.etsu.edu

Received 8 February 2006; Accepted 1 May 2006

DOI 10.1002/pbc.20913

While there have been a number of studies using qualitative methods to capture the experiences of the healthy siblings of pediatric cancer patients [14,15,17,19,26,27,29], only a few investigators obtained direct interviews with the siblings themselves [14,15,19]. Furthermore, many of these studies utilized mixed methods and focused specifically on adaptation, stress, and coping, instead of taking an inductive approach [17,19,26]. Existing data suggest that siblings are the ''forgotten family members'' during serious childhood illness [34]. Pediatric health care professionals are in a key position to identify and address the needs of these individuals, in order to help them adjust emotionally and to facilitate their normal development.

METHODS

Our study was a qualitative, descriptive analysis of the experiences of siblings of patients with FA. The goal of qualitative descriptive research is to produce ''a comprehensive summary of events in the everyday terms of those events'' [35]. In addition, qualitative description allows the reader to understand the essence of what it is like to experience a particular life situation. We used in-depth interviewing to identify and understand various themes which characterized the experience of living in a family with FA. Qualitative methods are a widely-accepted analytic strategy in the context of psychosocial research questions for which there is little or no information; in many cases it provides the stimulus for future research [36]. A unique benefit to this approach is that qualitative description is intended to result in improved consensus among researchers because it is less interpretive or abstract than other qualitative methods such as phenomenology, grounded theory, or ethnography [35]. Since the level of data abstraction was minimal, qualitative description was therefore especially appropriate to explore this clinically relevant topic in order to achieve usefulness amongst an audience of health care practitioners.

Healthy Siblings

Healthy siblings were recruited from families enrolled in an actively-accruing National Cancer Institute (NCI) protocol (02-C-0052) entitled, ''Etiologic Investigation of Cancer Susceptibility in Bone Marrow Failure Syndromes,'' which includes a cohort of North American FA families [37]. The protocol for the sibling study was approved by the Institutional Review Boards of the NCI Center for Cancer Research and the University of Pennsylvania Office of Regulatory Affairs.

Individuals between the ages of 11 and 21 years with at least one biologically related living half or full sibling with FA were eligible for this study. Our reason for recruiting siblings with a brother or sister living with FA was to narrow the focus to the current experience rather than retrospective reports. The sample size was determined through continuous data analysis aimed at an end point of data saturation, (i.e., at which additional material failed to yield new insights and redundancy was reached). In qualitative studies, redundancy is generally achieved with a small number of informants, especially if the depth of information is considerable [38]. In this study, redundancy occurred with the seventh interview; the interviewer (SPH) conducted two additional interviews to assure saturation.

The study was conducted between May 2003 and October 2005. Nine of 12 contacted siblings accepted and comprised the final sample. Seven of the nine interviews occurred in the home communities of the participants; two interviews took place at the National Institutes of Health Clinical Center. The interviewer provided a brief introduction, reviewed the purpose of the study, and obtained written informed consent from the siblings who were age 18 or older, and from the parents of those below age 18, and assent from the siblings between ages 11 and 17. A semi-structured guide facilitated the interview process, and was modified to include more focused questions as new information emerged during data collection. The interviewer developed the guide in conjunction with experts in pediatrics in order to structure the questions around the cognitive developmental stage of the informants [39]. The interview commenced with questions such as, ''Tell me a little bit about yourself'' and progressed to questions such as, ''What does Fanconi Anemia mean to you? And ''What is it like to have a brother or sister with Fanconi Anemia?'' Interviews lasted between 45 and 120 min and were audio-recorded and transcribed verbatim. The interview started with rapport-building questions, moved to open-ended questions regarding siblings' experiences, and then progressed to specific probes if specific discussion was not forthcoming.

The interviewer recorded field notes containing thick descriptions of the participants' behaviors, verbal and non-verbal cues, and environmental surroundings. She created case summaries (for within and cross-case analysis) to highlight the most relevant themes from each interview, and kept an introspective journal to note potential insights and to record contextual observations, such as social and environmental issues. These additional documents served as the study's audit trail and comprised a key part of the analysis.

Qualitative content analysis was used to inductively analyze interview data. In qualitative content analysis, codes which represent specific features of the data are derived from both verbal and visual data, in a real-time, ongoing process. The goal of the analysis was to extract themes from the observation of verbal and non-verbal data cues aimed at summarizing the most informational contents of the data [35]. The interviewer read and coded transcribed interviews using an iterative process, which involved highlighting and defining narrative units of text and organizing those units into descriptive schemes or categories. Ultimately, categories were grouped and elevated to a thematic level. The interviewer's doctoral dissertation committee provided

skeptical peer review and objective checks of confirmability (agreement between two or more independent people about the data's relevance) during the analytic process. These individuals, acknowledged herein, have profound expertise in qualitative methods. Transcript data were managed using NVivo 2.0 Software [40]. Overall, 1542 verbatim passages from the interviews were classified using 258 data-driven codes, and then categorized into four major themes, which will be detailed here.

Due to the rarity of FA and the small numbers of participants, the obligation to protect participants' confidentiality and anonymity prevents description of individual subject profiles. For this reason, references related to the gender of some participants were changed. Selected quotations from participants will be presented to illustrate the major themes.

RESULTS

Nine siblings (four female and five male) from seven families comprised the final sample. The median and range of ages of the informants were 13 and 11–21 years. The majority of the families identified themselves as white, non-Hispanic. The biological parents in six of the families were married. It is important to note that the physical appearance of the seven probands with FA was generally less severe than in many other patients with FA [41]. Two of the probands with FA had undergone a successful sibling matched bone marrow transplant (the siblings were participants in this study); another proband had a successful alternative donor transplant. Undergoing transplant was an imminent consideration for two additional probands at the time of the study.

Four major themes of the sibling experience were derived inductively from the interview data: (1) containment; (2) invisibility; (3) worry, and (4) despair. These themes are not mutually exclusive categories, nor can they be combined to form a more comprehensive construct. They interact and overlap, and together, characterize the sibling experience of living in a family with FA. Tables I–IV present selected quotes that are representative of each of the four main themes characterizing siblings' experiences. The themes are described below, along with the specific components within each theme.

Containment

"Containment" is the term we applied to siblings' and family members' tendency to set boundaries around, or confine, emotions and information related to FA. Siblings reported attempting to isolate FA from their day-to-day lives in order to limit its adverse impact. Siblings also reported parental unwillingness to engage in discussions related to the disease. This two-way conspiracy of silence exemplifies the containment theme. These components of sibling constraint and parental silence were frequently encountered (Table I).

Sibling constraint. Sibling constraint included the siblings' perceptions of the meaning of FA, as well as its impact on family life, and provided a window into their unspoken feelings about the disease. The participants related a symbolic representation of FA as something confining to them, illustrating their feelings of isolation, separation, and loneliness. Respondents also described investing enormous energy to keep their own distress under wraps, so as to avoid causing the family more worry or pain. The second passage in Table I depicts the deep emotional consequences of feeling isolated because "nobody understands."

Parental silence. Parental silence was a component that the majority of siblings reported in their accounts of how they initially learned (usually indirectly) about FA. Participants implied that their parents could not be approached directly to inquire about FA; their curiosity forced them to find other sources of information.

Invisibility

"Invisibility" is another prominent theme of the siblings' experience with FA. It captures experiences reported by respondents that left them feeling overlooked, and excluded. Siblings primarily spoke indirectly about their feelings of invisibility, as illustrated by the material in Table II.

TABLE I. Containment

Components	Illustrative quotes[a,b]
Sibling constraint	Everyone is born with a mixed bag of [sic] Pandora's Box, if you will. You've just got to make sure you keep the snap of that lid shut . . . guess that's your only option, all this vile [stuff] is coming out of the box and you've got to deal with it [16–21].
	It's the type of thing with FA you feel like you can't talk to anyone about it. 'Cause you feel like people don't get it. [You ask others to] think of leukemia, now think of [barely anybody] having it, and it being worse, and nobody understands . . . that's what I am dealing with. I have been face to face with death since [a young age], and that changes people [11–15].
Parental silence	Well, I was 5 years old I think, and I just [over]heard it from my mom and dad talking . . . sometimes I was eager to look in the mail to see if the words [Fanconi's Anemia] came up again. If there was an envelope with the words, I would open it [11–15].

[a]Illustrative quotes have been slightly paraphrased to increase readability.
[b]Following each quotation is a general age range of the participant from whom the passage was taken.

TABLE II. Invisibility

Components	Illustrative quotes[a,b]
Staying under the radar	My mom and I tend to fight when we start talking about FA stuff just because we don't communicate really well. So, I don't know, I try not to fight. It's easy for our discussions to get off track, and then there will be a fight and we just feel bad about it. It's tough [16–21].
	I try to make my mom happy by getting good grades in school [11–15].
Caretaking of family members	I do lots of chores for my mom and chores for my brother because he really doesn't feel good all the time [11–15].
	[My mom] will be crying and I will go over and comfort her and tell her you can't really do anything about it. So there is no use crying about things you can't change. So I will tell her that [16–21].
Being busy with patient-related activities	[I'm not] able to do a lot of stuff. I used to be able to just go out and have fun and now, since we have things to do and appointments, I can't really go out that much. That makes me mad because I like spending time outside with my friends; and then, whenever I do go somewhere, I get into trouble for staying out too late [16–21].
	We have a lot more things to do now. We are really busy; we barely ever get to just sit at home and be free for a day [11–15].

[a]Illustrative quotes have been slightly paraphrased to increase readability.
[b]Following each quotation is a general age range of the participant from whom the passage was taken.

Staying under the radar. Staying under the radar was a people-pleasing component that siblings used to avoid conflict and gain approval. They acknowledged the influence of their parents' expectations when talking about behavior and academic achievement. Siblings reported wanting to alleviate their parents' sorrow and worries related to the demands of caring for their sick brother or sister.

Caretaking of family members. Caretaking of family members was another manifestation of participants subordinating their own needs and desires, by focusing time and energy on the care and protection of others. They movingly spoke of how difficult life was for their parents and for their ill sibling, but did not seem to recognize their own needs in that regard.

Being busy with patient-related activities. Being busy with patient-related activities is another component of invisibility in which siblings were included in health care activities related to the child with FA. Siblings indicated that they rarely had an opportunity to take a break from thinking about FA. In some situations, this busyness caused siblings to disappear from their social milieu.

Worry

The third major theme was that of ''worry.'' Siblings were asked to describe what they worried about. While they identified a few age-appropriate worries, such as concern over friends or school, most of their worries centered on FA and its effect on their family and the future (Table III).

Prognosis for the affected sibling. Prognosis for the affected sibling was the most common worry cited by respondents. In most cases, siblings recognized that there was a chance that their brother or sister would die. For families in which bone marrow transplant was a consideration, much of the siblings' worries centered on the risks involved with the procedure and the potential of cancer following transplant.

TABLE III. Worry

Components	Illustrative quotes[a,b]
Prognosis for the affected sibling	I guess I worry that someday she'll (my sister) go out and a friend might bring her somewhere, and she'll be really close to a [dangerous] chemical or fall down, and break a couple bones. So that's my pretty much greatest fear . . . I think I'll be at my sister's funeral, and I think it will be too soon [16–21].
	I heard my mom and dad talking about a 4 out of 6 [match for transplant] and I know that there is a risk of it not working. I am kind of worried about taking that chance because my sister might die [11–15].
Transmission of FA to offspring	Yeah, I always thought that if I had it, I would reconsider having my own kids. I don't want the risk of having them die. And so, I would have a few of my kids and then adopt some [11–15].
Stigma of being in a family with FA	I'm worried about what my friends will say about my family. I worry that some of them are acting really strange. You worry if they are going to reject you, or like ignore you, for a long period of time [11–15].

[a]Illustrative quotes have been slightly paraphrased to increase readability.
[b]Following each quotation is a general age range of the participant from whom the passage was taken.

TABLE IV. Despair

Components	Illustrative quotes[a,b]
Sadness	Well, sometimes we are happy because nothing goes wrong. And sometimes [my sister] doesn't go to the hospital for a while, because she doesn't need platelets or blood or white cells. Sometimes we are happy, but that is kind of for a short period of time, because she always gets a chemical reaction. It's kind of sad. (Tearful) [11–15].
	You just try to have to learn to deal with it (FA). It's like the promise of suffering. You got to suffer. Suffer a lot and if you're not, you're not paying attention [16–21].
Jealousy	I was [ticked] off and jealous. She had something I didn't. She got attention. Maybe not so much jealous, as ''Yeah, give her attention but what about me?'' But I guess it really wasn't my time for attention [11–15].
Loneliness and Abandonment	I kind of felt like I fell through the cracks a little bit, for a little while. I think I was in some ways left to fend for myself [16–21].
Uncertainty	That's kind of like her course for that day, so far, so good; so far so good, 'cause you know you're going to hit the ground, eventually. You don't know how you are going to hit it, what position your body is going to be in, but you're going to hit the ground. I am sure [my sister] knows that. I'm sure it really [messes] her up [16–21].

[a]Illustrative quotes have been slightly paraphrased to increase readability.
[b]Following each quotation is a general age range of the participant from whom the passage was taken.

Transmission of FA to offspring. Transmission of FA to offspring was a concern that siblings cited when discussing their own future children. The passage presented in Table III demonstrates the sibling's confusion and worry about the differences between being a carrier versus being affected, and also about the implications derived from the autosomal recessive nature of FA.

Stigma of being in a family with FA. Stigma of being in a family with FA was another worry that siblings discussed. Their feelings of rejection were further compounded by the inability to talk about FA with their friends. Other siblings discussed how financial difficulties in their family (often a result of overwhelming medical expenses) caused embarrassment at school. With FA overshadowing their existence at home, one might hope that siblings' peer groups would be an important source of escape and support, but their words suggest otherwise.

Despair

''Despair'' represented the fourth major theme of siblings' experience. This theme incorporates elements of sadness, jealousy, loneliness and abandonment, and uncertainty (Table IV).

Sadness. Sadness was one of the most frequent emotional responses to having a sibling with FA. Generally, siblings did not describe their sadness explicitly, but it was implicit in much of what they said. Several of the siblings cried during the interview. Their persistent sadness appeared to engender a sense of helplessness.

Jealousy. Jealousy was created by the level of attention that the affected brother or sister received once the diagnosis of FA had been made. Overall, most siblings recognized that their parents were diverting their attention to the ill child for a ''good reason,'' and claimed that their jealousy was episodic and transient. Conversely, two of the respondents felt that their affected sibling was still getting ''special treatment'' despite doing well physically and having been diagnosed years ago.

Loneliness and abandonment. Loneliness and abandonment were not explicit themes, but these emotional sentiments nonetheless seemed to contribute to siblings' feelings of invisibility. Despite recognizing their parents' efforts, siblings felt marginalized nonetheless.

Uncertainty. Uncertainty was another recurring issue in siblings' experience. The vague and sometimes insidious onset of FA, its variable and unpredictable course, as well as the lack of reliable prognostic indicators, may all have contributed to siblings' uncertainty about what the future might hold for their affected brother or sister, their family unit, and themselves.

DISCUSSION

Chronic childhood illness pervades every aspect of family life, including the psychosocial well-being of healthy children; unaffected siblings experience significant emotional sequelae as a consequence of their brother or sisters' illness. Most notably, our study participants reported:

Investing enormous energy in trying to keep their own distress ''contained'' to avoid causing the family more worry or pain;
Feeling invisible, an experience which compromised their own sense of self or identity;
Assuming a caretaking role towards the parents and their affected sibling;
Having their normal routine disrupted by accompanying the affected child to doctors' appointments and hospitalizations;
Worrying about the prognosis of the affected child;
Experiencing despair, including sadness, jealousy, loneliness/abandonment, and uncertainty.

Overall, the lives of siblings seemed tied to what they thought was in the best interest of the child with FA. The experiences reported above provide evidence of a much broader array of psychosocial issues than one might have anticipated in children designated as ''unaffected'' by FA. While siblings do not have the FA phenotype, the impact of this disease on their individual lives is profound and may place them at increased risk of various psychopathologies, such as depression, anxiety, and post-traumatic stress [24,42,43].

Experience is a multi-dimensional construct and while the four major themes are not all-inclusive, they provide a starting framework for approaching healthy siblings in the clinical setting. While supporting siblings as they struggle to meet their own developmental goals and find a meaningful place for themselves in a family undergoing long-term illness is not an easy task, it is not insurmountable. The first step in addressing these issues in the clinical setting is to recognize that serious problems may exist among family members who are generally regarded as well. Parental observations are unlikely to provide a sufficient basis for intervening, as parents may not accurately perceive the problems of their healthy children. Although siblings may appear to be fine to the outside world, our data suggest that they are paying an emotional price as a consequence of their family situation. This host of challenges cannot help but raise serious concerns regarding siblings' long-term emotional and mental health.

Our findings suggest that the communication patterns of the siblings may be critical indicators of their isolation and invisibility. Siblings in this study avoided talking about FA within the family to protect family members from feeling sad and avoided talking about FA with individuals outside the family because they saw no way to make others understand. In fact, the interview for this study was often the first opportunity the siblings had to be the focus of attention within the health care system. This occurred across the entire age range of informants. Their circumstances have made these youngsters wise beyond their years, and remarkably articulate in relating their concerns. Providing them with formal opportunities to discuss their feelings (e.g., yearly physical exams and support groups) may result in improved adjustment to the illness and family life.

Various investigators have examined aspects of the psychosocial impact of chronic illness on healthy siblings. Our findings are congruent with this literature to the extent that it is possible to make comparisons between a complex, multi-system genetic cancer-predisposition disorder such as FA, and the conditions studied by others (e.g. cancer, sickle cell disease, cystic fibrosis). Stress and distress are common in siblings of children with chronic illness. Thus, our findings cannot be interpreted as ''abnormal'' experiences. Instead, these findings should be conceptualized on a variable continuum, whereby siblings are adjusting to the stress of chronic illness in what may be an adequate manner. Very little is known, however about the long-term adaptation of siblings of children with chronic illness. The results of a recent study exploring psychological adaptation in siblings of pediatric cancer patients suggested that emotional distress may continue well beyond the diagnosis of cancer in the affected child [32].

Our study extends the existing literature by identifying the specific dimensions of the unaffected sibling's experiences in an FA family. Our findings have important implications for understanding other rare genetic disorders, such as those which are part of the family of inherited bone marrow failure syndromes. Specifically, this study identified the theme of containment as a unique theme which encompassed the idea that both siblings and parents contained their feelings about FA. It is possible that over time this pattern of closed communication (containment on both sides) between family members may have serious psychosocial consequences for siblings such as feelings of resentment and anger toward the ill child or the parents as well as major implications for future relationships with family members.

This study also identified siblings' worries about themselves or future children with regard to FA. Based on the recessive pattern of inheritance, siblings shared fears that they might be at risk for developing FA or eventually passing it on to their children. Some siblings remarked that they might refrain from having their own children. Thus, this finding adds to existing knowledge by enhancing health care providers' and parents' understanding that children and young adults contemplate reproductive decisions well before what may be expected.

This study was limited by insufficient heterogeneity of the participants. For example, the sample did not include participants whose siblings had died from complications of FA, nor did it include siblings who had donated bone marrow to a sibling with FA who then died. In addition, the somewhat mild physical appearance of the probands meant that the presence of disease was not always apparent to individuals unfamiliar with the patient. Conceivably, siblings whose brother or sister had more obvious syndrome-related physical abnormalities may have experienced stronger negative reactions from persons outside the family and possibly even more distressing experiences. Alternatively, the lack of physical features may have caused the sibling to feel more confused since affected brothers and sisters did not often appear to be very ill.

The study sample was recruited entirely from those families who were voluntarily participated in an NCI clinical research protocol. They were primarily middle class, English-speaking, white individuals from well-educated families, with high motivation for participation in clinical research. Thus, although the NCI protocol is open to all FA families in North America, the subjects who actually participated may not be typical of all FA families. It is likely that there are additional unique experiences among those

whom we could not interview, since our sample was one of convenience.

Despite these limitations, the importance of the study results suggest that the sibling experience warrants further investigation. Considerations for future research include: (1) expanding the study size and participant diversity to further explore the experiences of siblings; (2) creating interventions to promote healthy adaptation to the illness within the family; and (3) exploring the experiences of siblings of patients with other chronic and/or genetic diseases.

Our data suggest that unrecognized psychosocial issues exist for the apparently healthy siblings of patients with FA. These findings support the need for an increased awareness among health care providers and parents as well as hypothesis-driven investigation, and the development and testing of assessment instruments to facilitate identification of problems with potential psychological morbidity.

ACKNOWLEDGMENT

The authors thank the participants and their families. S.P.H. also expresses gratitude to Drs. Mark H. Greene, Deborah B. McGuire, Janet A. Deatrick, and Frances K. Barg for their generous assistance as thesis advisors for this research. We additionally thank Dr. Mark H. Greene, Dr. Deborah B. McGuire, Dr. Robert W. Miller, and Jennifer T. Loud, RN, CRNP for critiquing and editing portions of this manuscript.

REFERENCES

1. Alter BP. Inherited bone marrow failure syndromes. In: Nathan DG, Orkin SH, Look AT, Ginsburg D, editors. Nathan and Oski's hematology of infancy and childhood, 6th ed. Philadelphia: W.B. Saunders; 2003.
2. Azarnoff P. Parents and siblings of pediatric patients. Curr Probl Pediatr 1984;14:1–40.
3. Cicirelli VG. Feelings of attachment to siblings and well-being in later life. Psychology and Aging 4:211–216.
4. Cicirelli VG. Sibling relationships in adulthood. In: Pfeifer SP, Sussman MB, editors. New York: Harworth Press. 1991;16:291–310.
5. Pulakos J. Young adult relationships: Siblings and friends. J Psychol 1988;123:237–244.
6. Fleitas J. When Jack fell down . . . Jill came tumbling after. Siblings in the web of illness and disability. MCN Am J Matern Child Nurs 2000;25:267–273.
7. Gogan J, Koocher G, Foster D, et al. Impact of childhood cancer on siblings. Health Soc Work 1977;2:41–57.
8. Harding R. Children with cancer: The needs of siblings. Prof Nurse 1996;11:588–590.
9. Houtzager B, Grootenhuis M, Last B. Supportive groups for siblings of pediatric oncology patients: Impact on anxiety. Psycho-oncology 2001;10:315–324.
10. Koch-Hattem A. Siblings' experience of pediatric cancer: Interviews with children. Health Soc Work 1986;11:107–117.
11. Murray JS. The lived experience of childhood cancer: One sibling's perspective. Issues Compr Pediatr Nurs 1998;21:217–227.
12. Sloper P. Experiences and support needs of siblings of children with cancer. Health Soc Care Community 2000;8:298–306.
13. Sargent JR, Sahler OJZ, Roghmann KJ, et al. Sibling adaptation to childhood cancer collaborative study: Siblings' perceptions of the cancer experience. J Pediatr Psychol 1995;20:151–164.
14. Kramer RF. Living With Childhood Cancer: Impact on The Healthy Siblings. Oncol Nurs Forum 1984;11:44–51.
15. Chesler MA. Voices from the Margin of the Family: Siblings of Children With Cancer. J Psychosoc Oncol 1991;9:19–41.
16. Horwitz WA. Family adaptation to childhood cancer: Sibling and family systems variables. J Clin Child Psychol 1990;19:221–228.
17. Barbarin OA. Sibling adaptation to childhood cancer collaborative study: Parental views of pre and postdiagnosis adjustment of siblings of children with cancer. J Psychosoc Oncol 1995;13:1–20.
18. Cowen L. Psychologic adjustment of the family with a member who has cystic fibrosis. Pediatrics 1986;77:745–753.
19. Walker CL. Stress and coping in siblings of childhood cancer patients. Nurs Res 1988;37:208–212.
20. Faux SA. Siblings of children with chronic physical and cognitive disabilities. J Pediatr Nurs 1993;8:305–317.
21. Heffernan SM. Behavior changes exhibited by siblings of pediatric oncology patients: A comparison between maternal and sibling descriptions. J Pediatr Oncol Nurs 1997;14:3–14.
22. Sargent JR. Sibling adaptation to childhood cancer collaborative study: Sibling's perceptions of the cancer experience. J Pediatr Psychol 1995;20:151–164.
23. Harder L. Siblings of children with cystic fibrosis: Perceptions of the impact of the disease. CHC 1982;10:116–121.
24. Alderfer MA, Labay LE, Kazak AE. Brief report: Does post-traumatic stress apply to siblings of childhood cancer survivors? J Pediatr Psychol 2003;28:281–286.
25. Sloper P. Risk factors in the adjustment of siblings of children with cancer. J Child Psychol Psychiatry 1996;37:597–607.
26. Sahler OJ. Sibling adaptation to childhood cancer collaborative study: Prevalence of sibling distress and definition of adaptation levels. Dev Behav Pediatr 1994;15:353–366.
27. Schuler D. Psychosocial problems in families of a child with cancer. Med Pediatr Oncol 1985;13:173–179.
28. Cohen DS. Pediatric cancer: Predicting sibling adjustment. J Clin Psychol 1994;50:303–319.
29. Lehna CR. A childhood cancer sibling's oral history. J Pediatr Oncol Nurs 1998;15:163–171.
30. Murray JS. Social support for school-aged siblings of children with cancer: A comparison between parent and sibling perceptions. J Pediatr Oncol Nurs 2001;18:90–104.
31. Murray JS. Self-concept of siblings of children with cancer. Issues Compr Pediatr Nurs 2001;24:85–94.
32. Houtzager BA, Grootenhuis MA, Caron HN, et al. Quality of life and psychological adaptation in siblings of paediatric cancer patients, 2 years after diagnosis. Psychooncology 2004;13:499–511.
33. Houtzager BA, Grootenhuis MA, Hoekstra-Weebers JE, et al. One month after diagnosis: Quality of life, coping and previous functioning in siblings of children with cancer. Child Care Health Dev 2005;31:75–87.
34. Van Dongen-Melman JE, De Groot FAU, De Groot AF, et al. Siblings of childhood cancer survivors: How does this ''forgotten'' group of children adjust after cessation of successful cancer treatment? Eur J Cancer 1995;31A:2277–2283.
35. Sandelowski M. Whatever happened to qualitative description? Res Nurs Health 2000;23:334–340.
36. Kearney MH. Levels and applications of qualitative research evidence. Res Nurs Health 2001;24:145–153.
37. http://www.marrowfailure.cancer.gov. World Wide Web. 10-31-2005. Ref Type: Electronic Citation.

38. Polit DF, Hungler BP. Nursing research: Principles and methods, 6th ed. Philadelphia: Lippincott Williams & Wilkins; 1999.
39. Deatrick J, Faux SA. Conducting qualitative studies with children and adolescents. In: Morse JM, editor. Qualitative nursing research: A contemporary dialgue. Newbury Park: Sage Publications; 1991. pp 203–223.
40. http://www.qsrinternational.com. World Wide Web. 2005. Ref Type: Electronic Citation.
41. Rosenberg PS, Huang Y, Alter BP. Individualized risks of first adverse events in patients with Fanconi anemia. Blood 2004;104: 350–355.
42. Cadman D. The Ontario Child Health Study: Social Adjustment and Mental Health of Siblings of Children With Chronic Health Problems. Dev Behav Pediatr 1988;9:117–121.
43. Breslau N. Siblings of disabled children: Effects of chronic stress in the family. Arch General Psychiatry 1987;44:1040–1046.